A CASE OF MULTIPLE LESIONS OF THE SPINAL CORD AND CRANIAL NERVES WITH AMYOTROPHY, DUE PROBABLY TO SYPHILITIC INFECTION.[1]

By Max H. Bochroch, M.D.,

CHIEF CLINICAL ASSISTANT IN THE NEUROLOGICAL DEPARTMENT AND
INSTRUCTOR IN ELECTRO-THERAPEUTICS IN JEFFERSON MEDICAL
COLLEGE; NEUROLOGIST TO THE OUT-PATIENT DEPARTMENT
OF ST. JOSEPH'S HOSPITAL;

AND

Alfred Gordon, M.D.,

ASSISTANT IN NEUROLOGY, JEFFERSON MEDICAL COLLEGE.

J. B., aged 34, brewer by occupation, came with the following history to the Neurological Clinic of the Jefferson College Hospital. In July, 1900, he began to feel pain in the right supra-orbital region, which lasted until October. Glasses were adjusted and pain became easier. In September a ptosis of the right eye developed very rapidly. At that time he noticed that he saw double, which condition has continued up to the present time. Two months later he began to complain of pain and weakness in the left thigh when he walked or stood, but when resting there was no pain. As soon as he starts to walk, pain appears in the left knee, and while walking extends to the whole thigh. In May, for the first time, he experienced shooting pain all along the left limb. Besides the pain the patient complained also of sensation of heaviness in the left thigh when he walked or attempted to get up, or on going upstairs. This condition of heaviness has been present from the onset of the disease. At about the same time he commenced to have some trouble with micturition: it was somewhat difficult; he had to wait a few minutes before he could urinate. This condition exists at the present time. He has been constipated for many years. Patient was always healthy. Never had any infectious disease, except gonorrhea about seven years ago; it lasted but eight days. He has been married for the last two years, has one child, which is healthy. No history of miscarriages. Has one brother of forty-two, who is in good health, two sisters who are married and in perfect health. Father and mother are about seventy and in good health. Patient had worked in a brewery since the age of fourteen, has therefore been exposed to cold and dampness. Alcoholism and syphilitic infection denied.

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[1] Read before the Philadelphia Neurological Society.

Present Condition.—Patient weighs 158 pounds, height 5 feet, 10 inches; chest circumference 41 inches. He is very well built and powerful. Excellent stature. Excellent and healthy appearance.

Examination shows: Gait normal with open eyes, but somewhat difficult with closed eyes. Cannot stand on the left leg, but does it with difficulty on the right. Station with closed eyes presents a normal sway. Sensations (touch, pain and temperature) normal. Besides heaviness he complains of cramp-like sensation in the left leg, occasionally in right. Knee-jerk on the right is exaggerated, on the left lost, even on reinforcement. Some rigidity in the right leg, flaccidity in the left. No ankle-clonus on either side. Babinski present on both sides, more marked on the left than on the right. Musculature: Upper extremities are apparently equally developed on both sides, no evident atrophy. The left scapula distinctly recedes from the thorax, and Bechterew's reflex is more marked on the left than on the right side. Dynamometer shows 75 for R. and L.

Lower extremities: Cyanosis on the left side, skin colder on the left than on the right. The vastus internus on the left is very flabby. The measurements are as follows: taken in the middle at equal distance from the upper border of the patella the circumference of the left thigh is 38 cm., of the right 44 cm. The legs at the same distance from the lower border of the patella show 27 1-2 cm., for the left leg, and 29 for the right. Left knee-joint is larger than the right. The musculature of the legs is too small in comparison with the rest of the body above the umbilicus.

Electrical examination gave the following result: *Left limb.* Examined with a galvanic current, the muscles of the whole member show that the AnCC is equal to the CaCC, more marked in the vastus internus, except that of the postero-external aspect of the thigh, where the CaCC is but very slightly stronger than AnCC. With a faradic current very little response to a current of average strength was found, and to obtain a normal contraction a very strong current was required. *Right limb.* Under galvanic current the muscles of the thigh present the normal formula, but those of the leg present some change: the CaCC is but slightly stronger than AnCC. *Eyes.* Right pupil larger than left, and does not react to light. Left reacts sluggishly to light. The right eye presents an ophthalmoplegia; this is very marked in the upper and lower movements of the eye; on the nasal side there is only a partial paralysis of the muscle. The movement of the eye-ball externally is complete. Ptosis was very marked when we first saw him, but it is improved. The ciliary muscles are also paralyzed.

Summing up the case we see a progressive muscular atrophy of spinal type limited to the lower extremities, pronounced in the whole left limb, and less marked in the right leg, with an increased deep reflex and rigidity on the right side, and loss of reflex with flaccidity on the left, slight disturbance of micturition—all this developed in a patient who presents a characteristic nuclear ophthalmoplegia. The case seemed to us quite unusual, and we hesitated in classifying it. The diagnosis can be made in our opinion only by exclusion. Multiple neuritis, tabes, anterior poliomyelitis of adults, amyotrophic lateral sclerosis and syphilis of the cerebro-spinal axis were taken into consideration.

Multiple neuritis may follow exposure to cold such as was the case in this instance, also give rise to loss of power early in the disease; the muscles atrophy rapidly and become flabby, the deep reflexes are lost. In spite of the presence of these few symptoms, we cannot accept the diagnosis of multiple neuritis in our case, as the onset, course and other symptoms are not to be found in this disease: we do not find the sudden onset of infectious character: no chills, no elevation of temperature, no premonitory symptoms, like malaise, headache, etc. Nor do we find the typical course of multiple neuritis. Tenderness of the nerve trunks under pressure is absent and the electrical reactions show changes which we do not find in multiple neuritis.

Anterior poliomyelitis should next be taken into consideration. This disease may occur in adults as well as in children. It is the result of an infection which has its seat of predilection in the gray matter of the anterior horns. If the acute form of this disease presents an onset infectious in character, there are, nevertheless, subacute and especially chronic varieties where the onset is slow, and the last form we find mostly in adults. In a few isolated cases of chronic poliomyelitis progressive muscular atrophy was found, but this is quite rare. Dejerine[2] reports a similar case in a syphilitic patient, but the atrophy was more marked in the upper extremities than in the lower. Autopsy proved that it was a case of a simple chronic poliomyelitis; in the whole cervical region the cells of the anterior horns had disappeared and the anterior roots were very much altered. The white columns remained normal. In our case atrophy is present in the lower limbs, but the other symptoms are not those of anterior chronic poliomyelitis, such as was conceived by Charcot and Vulpian. It remains for us to consider the possibility of amyotrophic lateral sclerosis, or of a multiple disseminated syphilitic infection of the cerebro-spinal axis. The first of these two affections should be thought of at the first glance.

We have here progressive muscular atrophy of the spinal type; gradual wasting of muscles of the whole left extremity, and partially of the right leg; the fibrillary contractions are absent, but the electric reactions are in accord with the idea of a spinal muscular atrophy; the knee-jerk is lost on the left side, but exaggerated on the right; flaccidity on the left, rigidity on the right side; Babinski's sign present on both sides. We may therefore admit an involvement of both the gray matter and lateral columns on the right side of the cord, but an involvement of only the gray matter on the left. Consequently we are in presence of multiple lesions distributed unequally on both halves of the cord, and the classical type of amyotrophic lateral sclerosis cannot be accepted.

Multiplicity of lesions all along the cerebro-spinal axis, manifold in their manifestations, various in localizations, supervening almost simultaneously in different portions of the cord, extending to the medulla and involving nuclei of cranial nerves, all this suggests syphilitic affection of the nervous system. The only unusual atypical point in our case is the amyotrophy.

Muscular atrophy of myelopathic type in systemic diseases of the cord is quite rare. Dejerine thought that tabetic amyotrophy was of peripheral nature, but Leyden, Charcot, Pierret, Marie have found alterations of the anterior horns. The microscopical studies show that the degeneration of the cells of the anterior horns may take place in two ways: one is the ordinary poliomyelitis, an acute disease which destroys not only the cells, but also other histological elements of the anterior horns; it is an affection of the trophic center which alters the peripheral motor path. The other is an affection of the anterior horns which begins in an insidious way and very slowly; we find it in toxic and tabetic amyotrophies. The clinical character of tabetic amyotrophy is totally in accord with the histological changes. We know that the collaterals of the posterior roots reach and surround the motor cells of the anterior horns. In tabetic degenerations those collaterals disappear, and if the stimuli for the motor and trophic cells of the cord no more exist, we can readily understand why we find the changes in those cells, changes that are first of dynamic and then histologic character. But if amyotrophies in tabes are rare, they are still rarer in specific affections of the cord. The perusal of the literature at our disposal for the last five years convinced us that there are only a few reported cases of amyotrophy in syphilis. The older writers mention occasionally syphilis as the causative factor of muscular atrophy. In one case Shmauss reports a flaccid paralysis and rapid wasting of the muscles, and at autopsy a focus of softening in the gray matter of the lumbar cord.

Among recent writers we find one case of Rendu and three cases of Raymond[3]. They all present a clear history of syphilis with amotrophy of Aran-Duchenne's type. Like in our case, the involvement began with a diplopia of the right eye. Discussing the case, Raymond arrives at the conclusion that it was a case of a chronic meningo-myelitis of a diffuse and progressive character. At autopsy he found, besides other lesions, an involvement of the anterior roots and these roots were thin and gray; and disease of the anterior horns where the cells were very much altered, were without prolongations, and without deformed nuclei. The cellular changes were only in the cervical region, corresponding to the amyotrophy of the upper extremities. But what was striking was the vascular involvement; the vessels were large, overfilled with blood; the same condition was seen in the anterior horns. The white columns were also very much altered. The meninges were involved; those of the spine were soft, and the dura of the cranium was adherent. As to the ravages of syphilis in other portions of the cord, they are too well known to dwell upon. The etiological relation between syphilis and various nervous diseases has been established comparatively recently.

This is particularly true with regard to the spinal cord. The history of Erb's spastic spinal paralysis, or of Charcot's spastic dorsal tabes illustrates sufficiently the rôle of specific infection. Baumgarten and Lanceraux maintain that the first stage of syphilis of the nervous centers is characterized by a periarteritis, which is soon followed by an endarteritis, because of a deficient nutrition caused by an obliteration of the vasa vasorum. The pia is the starting point; the arteries and arterioles which nourish the substance of the cord being obliterated, ischemia is produced, and the gray matter principally suffers from insufficient irrigation of blood. Ehrlich, Brieger and Spranck proved it experimentally. Similar results were obtained by Flourens[4] and by Panum[5]. If syphilitic infection is capable of injuring various portions of the cord through the large and smallest blood-vessels, the lesions will be various. The predominance of the lesion in certain tracts or in the gray matter will give a picture of systemic spinal disease.

Oppenheim, Ewald, Marinesco called attention to cases of spinal syphilis associated with symptoms of tabes. Minkowski observed a case in which the lesion was localized almost exclusively in the lateral columns, and gave the picture of a primary lateral sclerosis. Mendel and Schuster observed multiple specific lesions in the cord in a case which clinically presented the signs of disseminated sclerosis.

According as the process of destruction of the gray substance is rapid or slow we may observe the clinical types of acute or chronic anterior poliomyelitis.

Syphilis is capable of causing amyotrophic paralysis not only as a result of a myelitis or meningo-myelitis like in Raymond's cases, but also as a result of a general peripheral neuritis. Gilles de la Tourette's case is the best illustration of this.

Finally there are complex forms which cannot be classified, and in which we find symptoms of involvement of the roots, of the white columns and of the gray matter, symptoms of multiple and disseminated lesions along the spinal axis.

In a recent paper one of us[6] brought out the importance of infection as etiological factor in various affections of the cord with numerous clinical, pathological and experimental data to corroborate and emphasize this view.

In presenting this case we take the opportunity to again call attention to this point. In all probability our case is one of those many examples where a spinal amyotrophy was caused by infection. The previous history, the course of the disease, the fact that the patient could take high doses of specific remedies, and the amelioration of symptoms under this treatment are in favor of the infectious origin of the muscular atrophy.

We wish to express our indebtedness to Dr. F. X. Dercum for his kind permission to present this case.

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[2] Comp. Rend. de la Soc. de Biol. 95, 10, s II, 188.

[3] Bull. de la Soc. Méd. des Hôp., Février, 1893.

[4] Computes rend. de la Acad. de Sci., 1847.

[5] Exper. Beiträge zur Lehre von der Embolie. Virchow's Archiv., 1862, Bd. xxv.

[6] A. Gordon, Philadelphia Med. Jour., 6-29-1901.